Multiple Intracranial Aneurysms as Delayed Complication of Atrial Myxoma

Case Report and Literature Review

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Summary

We report a case of multiple intracranial aneurysms as delayed complication of atrial myxoma. We reviewed the literature of intracranial myxomal aneurysms, and trying to find reasonable therapy methods, but got the conclusion that neurosurgery and interventional treatment were not helpful, chemotherapy and radiotherapy maybe useful in the treatment of such cases.

Introduction

Myxomas are the most common benign tumors of the heart, accounting for approximately 50% of primary heart tumors. About 45% of left atria myxomas lead to systemic embolization, 50% of the accidents are cerebrovascular. Myxomas can also lead to intracranial aneurysms. To date there have been only 16 cases of cerebral aneurysm related to atrial myxoma reported in the literature.

The mechanism and the natural history of intracranial myxomal aneurysms are still controversial. Intracranial myxomal aneurysms are mostly distributed along the branches of the anterior and middle cerebral arteries, and the optimal treatment is in a dilemma. This prompted us to review the literature and to discuss the pathogenesis and optimal treatment of this disease.

Case Report

A 19-year-old female was transferred to our hospital, complaining of epileptic attacks on being excited. The seizures numbered about ten times a day and lasted about 2-3 minutes, without loss of consciousness; during the intermission of the attacks the patient was aphasic. The patient was admitted for symptomatic epilepsy. The patient had been diagnosed with left atrial myxoma, which was resected two years ago.

On admission the patient was conscious, with bradyphrasia and left central facial palsy. The muscle power of the right upper and lower limbs was 0/5 grade, the muscle tone was low, and the Babinski sign was positive. The MRI on admission showed a fresh infarction beside the right ventricle, cerebromalacia in the left basal ganglia, and a lacunar infarction in the right frontal subcortex (figure 1). MRA detected two regional protrusions on the left MCA, suspected to be aneurysms. TCD detected a 4.4-3.6 mm aneurysmal dilation on the crotch of M2 of the left MCA. Cerebrovascular DSA detected regional dilation on the distal end of the left MCA M1 segment (figure 2), and many saccular dilations on the distal end of the MCA and PCA of both sides; and visualization of the distal vessels was obviously retarded (figure 3). echocardiography did not detect a recurrence of the atrial myxoma. The patient was undergone anti-epi-

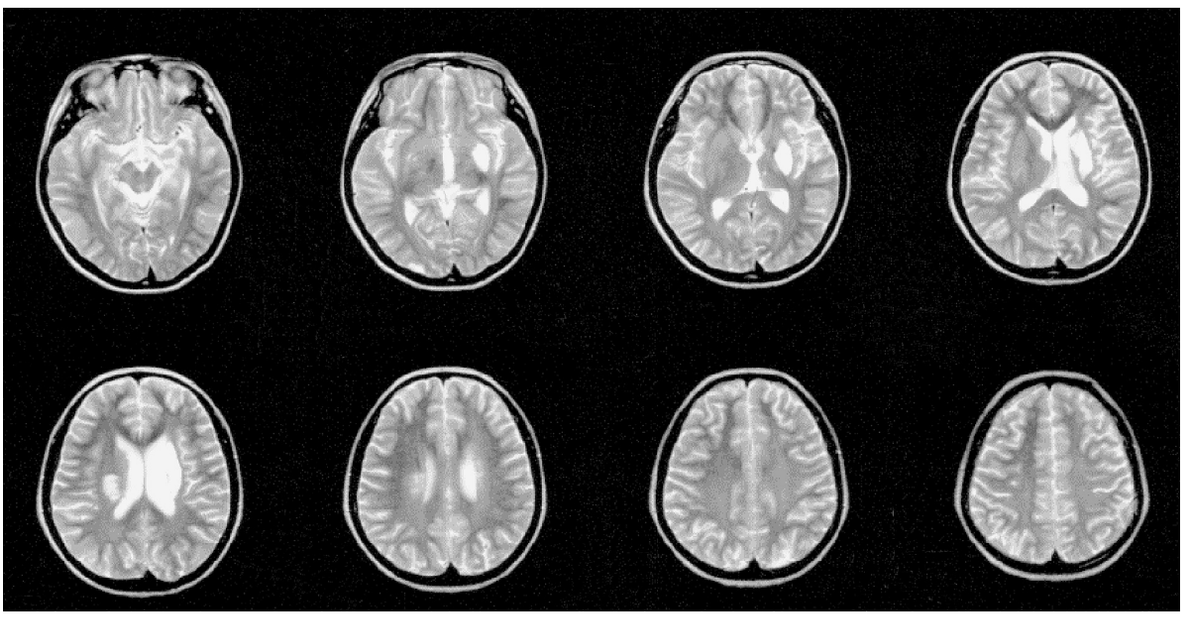

Figure 1 The MRI on admission showed fresh infarction beside the right ventricle, cerebromalacia in the left basal ganglia, and lacunar infarction in the right frontal subcortex.

leptic treatment, Sodium valproate 200mg was given orally every eight hours, and no epilepsy seizured any more. Further treatment included neurotrophic and rehabilitation of the affected limbs. One month later before discharged the muscle power of the patient's right limbs had recovered to 3/5 grade. The patient was followed up for one year with CT, no new ischemic foci or haemorrhagic foci was found.

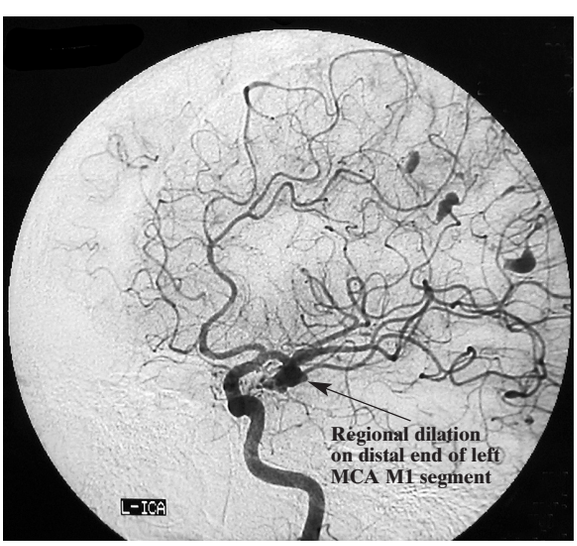

Figure 2 DSA detected regional dilation on distal end of left MCA M1 segment.

Discussion

Myxomas are the most common benign tumors of the heart, accounting for approximately 50% of primary heart tumors. It occurs sporadically and mostly afflicts 30 to 60-year-old females. Rarely, it presents as familial cases, or as Carneg's symptom complex, namely, myxoma accompanied with cutaneous, mammary gland and pituitary gland disorders [1]. Myxomas may occur in any heart chamber and attach on the endocardium, or even on the cusp of valves, 75% being in the left auricle, close to the fossa ovalis, 18%, 4%, 4% in the right auricle, right ventricle and left ventricle respectively. Most scholars think that the tumor cell is derived from the endothelial cell, and may be related to heterogeneity of mesenchyme cells and heteroplasia of embryonic rests [2]. Under electron microscopy, the myxoma cell is similar to embryonic rests of fossa ovalis, because tumors mainly originate there [3]. About 45% of left atria myxomas lead to systemic embolization, 50% of the accidents are cerebrovascular and may cause visual disorder, headache, atony of limbs and epilepsy. Repeated embolizations can lead to progressive dementia. The neurological symptoms secondary to myxomas can be acute or chronic. In the acute phase, tumor embolization results in ischemic brain disease due to

vascular stenosis and occlusion. Sandok reported that 25% of the patients was affected with neurological defects before being correctly diagnosed.

To date, several cases of delayed neurological dysfunction related to myxoma embolization have been reported. Saccular aneurysms and irregular arterial stenosis were found in imaging examinations. Intracranial myxomal aneurysms are mostly distributed along the branches of the anterior and middle cerebral arteries. Myxoma embolization can sometimes induce saccular aneurysms. Emboli arising from aneurysms can lead to transient ischemic attacks, while imaging examinations are mostly negative. Haemodynamic changes secondary to significant arterial stenosis cause transient ischemic attacks.

Metastasis to the cerebral and ventricles seldom occurs. Budzilovich reported one case of myxomal embolization in which the tumor grew through the vessel wall into the brain tissue, forming metastastic lesions and induced corresponding symptoms [6]. Delayed neurological symptoms are rare after myxoma resection. Knepper followed for eight years eight patients whose myxomas were resected, and did not detect any new neurological complications [7]. Sandok followed 35 cases of myxoma patients for four years after operation; only one patient developed cerebral ischemic symptoms [5].

There are two theories about the pathogenesis of myxomal aneurysms. The first theory proposes that vascular wall injury results from haemodynamic changes induced by embolization. The second theory argues that myxomal emboli can erode the vascular wall, a process which has been proved by pathological examination [8,9]. Myxomal emboli can form fusiform nests of tumor cells, which infiltrate into the vascular wall, and grow under the endothelium, thereby destroying the vascular wall and inducing fibrosis; the secondary inflammation and fibrosis of the elastic layer cause corresponding vascular pathological changes, such as stenosis or occlusion of the lumen, nodular thickening of vascular wall, and aneurysmal dilation. The process progresses slowly, so aneurysms may develop later several years after resection of the myxomas. To date there have been only 16 cases of cerebral aneurysm related to atrial myxoma reported in the literature [10].

The first clinical manifestation of the present patient was cerebral embolism, and epilepsy developed after myxomal resection.

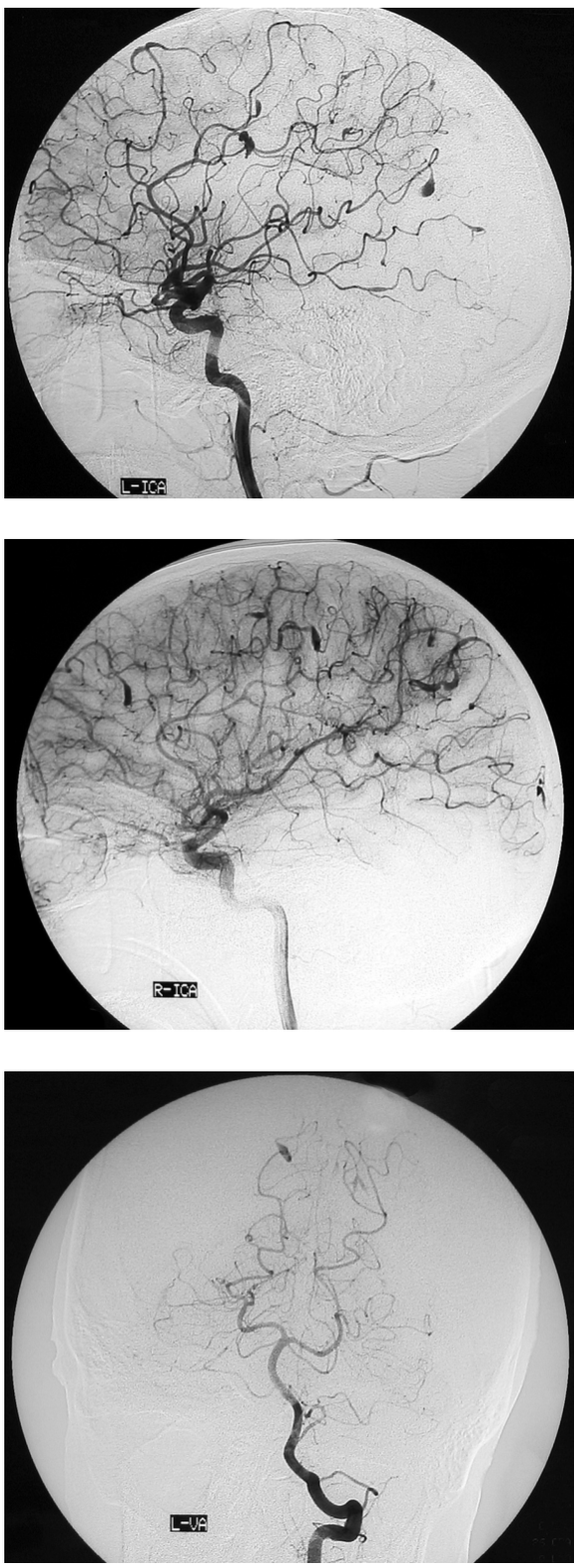

Figure 3 Saccular dilations on the distal end of the MCA and PCA of both sides and visualization of the distal vessels obviously retarded.

The seizures may be related to the paradoxical discharge of the cortex induced by the aneurysms. MRI and DSA detected multiple intracranial aneurysms, as in past medical reports. It is hard to explain why MRI detected a new infarction beside the right ventricle, for echocardiography did not detect a recurrence of the atrial myxoma, thus ruling out cardiogenic embolization. It is conjectured that the new infarction could be caused by the dislodgment of old emboli or by an unrelated local arterial disease. Another characteristic of this case is that the distal end of the left M1 segment was dilated like a saccular aneurysm. Most of the myxomal aneurysms reported in the literature were located on the small branches distal to the circle of Willis. This is one of the very few cases that an aneurysm was located on the stem of the MCA.

The natural history of myxomal aneurysms is not clear yet. Some scholars report that the aneurysms can enlarge or disappear in the course of the disease. There are also reports that myxomal aneurysms can rupture and cause subarachnoid haemorrhages or intracerebral haematomas. Bobo and Evans reported a patient with myxomal aneurysms causing left thalamic haemorrhage, but the aneurysms were on the distal part of the right MCA, so the relationship between the myxomal aneurysms and the intracranial haemorrhage was not clear [11].

Roeltgen applied adriamycin to treat one case of progressive myxomal aneurysms on the right MCA, and after six months the aneurysms stopped to enlarge [10]. The effects of chemotherapy and radiotherapy are uncertain, since the natural history of myxomal aneurysms is not well known. In our opinion since the myxoma cells implanting and penetrating the vessel wall have been proved by histological evidence [8,9], intravenous and intra-arterial chemotherapy may be most prospective therapy methods. Jean tried to isolate and resect the aneurysms located on the cortical arteries in a case of cardiac myxoma, but since they were multiple and many involved the terminal branches of numerous intracranial arteries, the operation failed to affect a complete cure. Further study should be performed to decide the merits of operation and endovascular treatment in treating myxomal aneurysms located on the main branches of the cerebral arteries. In the present case, the aneurysm was located on the distal end of the right MCA and its shape was irregular, so surgical treatment was not indicated.

References

1 Akabas H, Kirati K, Deglar B et Al: Surgical treatment of left atrial myxoma in Carney's complex. Thorac Cardiovasc Surg 45: 148-150, 1997.
2 Burke AP, Virmani R: Cardiac myxoma. A clinicopathologic study. Am J Clin Pathol 100: 671-680, 1993.
3 Kosuga T, Fukunaga S et Al: Surgery for primary cardiac tumors. Clinical experience and surgical results in 60 patients. J Cardiovac Surg (Torino) 43: 581-587, 2002.
4 Mattle HP, Maurer D, Sturzenegger M et Al: Cardiac myxomas: a long term study. J Neurol 242: 689-694, 1995.
5 Sandok BA, von Estorff I, Giuliani ER: Subsequent neurological events in patients with atrial myxoma. Ann Neurol 8: 305-307, 1980.
6 Budzilovich G, Aleksic S et Al: Malignant cardiac myxoma with cerebral metastases. Surg Neurol 11: 461-469, 1979.
7 Knepper LE, Biller J et Al: Neurologic manifestation of atrial myxoma: A 12-year experience and review. Stroke 19: 1435-1440, 1988.
8 Furuya K, Sasaki T et Al: Histologically verified cerebral aneurysm formation secondary to embolism from cardiac myxoma. J Neurosurg 83: 170-173, 1995.
9 New PF, Price DL, Carter B: Cerebral angiography in cardiac myxoma. Correlation of angiographic and histopathological findings. Radiology 96: 335-345, 1970.
10 Roeltgen DP, Weimer GR, Patterson LF: Delayed neurologic complications of left atrial myxoma. Neurology 31: 8-13, 1981.
11 Jean WC, Walski-Easton SM et Al: Multiple intracranial aneurysms as delayed complications of an atrial myxoma: case report. Neurosurgery 49: 200-202; discussion 202-203, 2001.
12 Bobo H, Evans OB: Intracranial aneurysms in a child with recurrent atrial myxoma. Pediatr Neurol 3: 230-232, 1987.

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